Progressive Facial Hemiatrophy with Some Unusual Symptoms

BY

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PROFESSOR OF DISEASES OF THE MIND AND NERVOUS SYSTEM IN THE NEW YORK POLYCLINIC

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Progressive Facial Hemiatrophy, with Some Unusual Symptoms.[1]

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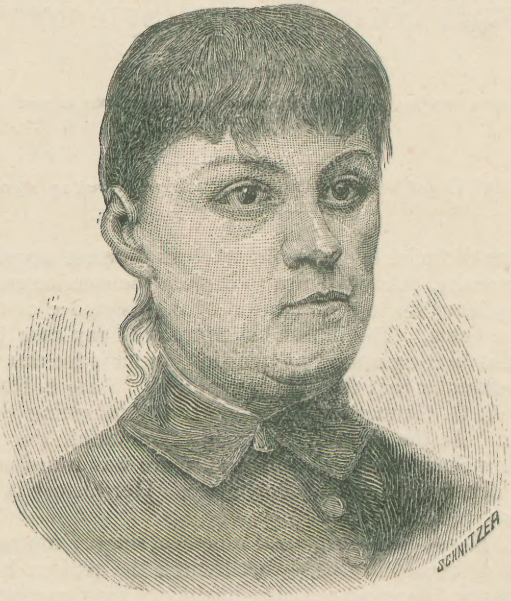
CASES of facial hemiatrophy have an interest apart from their rarity. Any attempt to explain the symptoms necessarily leads up to the question of the existence of trophic nerve-fibres. The unusual symptoms of my case have a special bearing upon this question, and it is for this reason in particular that the case is reported. I am indebted to Dr. A. G. Gerster for the opportunity of seeing this patient. At the time of the first examination she was in Mount Sinai Hospital, and from there was transferred to my department in the Polyclinic. Her history is as follows:

E. K——, Russian, aged nineteen, single; mother died from unknown causes, father living; the patient has one unmarried sister who is also healthy. First menstruation at the age of fifteen years. She was entirely well until about one year ago, when she noticed a peculiar appearance of the skin just below the left nostril, and was also struck with the fact that the face grew thinner below the left eye and above the left angle of the mouth. There is

[1] The patient was presented to the Neurological Section of the New York Academy of Medicine, December 6, 1889.

no history of any antecedent emotional excitement or injury; no erysipelas or other acute infectious disease preceded the onset of the present trouble.

Examination.—A healthy-looking, robust girl. Right half of face chubby, and all parts well developed. Distinct atrophy of middle and lower third of left side of



face; the atrophy involves the skin, fat, and also the muscles and bone of this side of the face. The masseters stand out very prominently, in part as a result of the atrophy of other muscles, and in part because they present one of the unusual symptoms. At intervals of a few seconds only, a series of *clonic and then tonic contractions of the temporal and masseter muscles* can be ob-

served. During this time it is impossible for the patient to open the jaw, and the face wears a very anxious expression. As the spasm extends to the tongue, and sometimes begins with the tongue, speech is impossible while these contractions last. These muscular spasms occur very frequently, and are most apt to follow upon excitement or exposure to cold. They are not only annoying, but painl ful, and were the cause of the patient's seeking medica- advice.

Left side, distance from middle of chin to angle of jaw, 11 ctm.; on right side, $12\frac{1}{8}$ ctm. Left side, from upper margin of naso-labial fold to middle of ear, $10\frac{3}{4}$ ctm.; on the right side, $11\frac{1}{2}$ ctm.

Movements of face and jaw: Muscles supplied by facial on both sides act normally except on left side, the attempt to puff out cheeks reveals atrophy of the parts and tissues involved.

Action of masseters entirely normal until spasms supervene; lateral movements of lower jaw also of normal extent.

Tongue: Distinct atrophy of left half—another "unusual symptom"—and of left floor of mouth under tongue.

Appearance of skin: No peculiarity except the "scar" on left upper lip, which is slightly pigmented, and which is evidently due to the disappearance of the fatty deposit in the skin; the skin is neither thickened nor glossy; the eyebrows and eyelashes are the same on both sides and entirely normal. No unilateral sweating to be observed even after the administration of a hypodermic injection of pilocarpine.[1]

Tactile pressure and temperature-sense entirely normal. Pain sense diminished a very little on left side.

Taste, smell, and hearing unobjectionable. All these special senses were most carefully tested.

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[1] One-eighth of a grain injected under the skin of the forearm produced slight and general sweating; a higher dosage was not attempted, as the patient was made exceedingly uncomfortable by the above dose.

Ocular movements and pupillary reflexes entirely normal. Faradic response of temporal and masseter muscles, of facial nerves and muscles altogether normal; naturally the movements of the right side of face appeared more vigorous.

The examination of the other parts of the body gave negative results, the only point worthy of notice being this, that the repeated thermometrical measurements of external ear showed a difference of one degree in favor of *right* ear.

The history shows that a number of the more serious symptoms observed in other cases—the enophthalmos, the falling out of the hair, and other dermal changes were either but slightly developed or entirely absent. The case was distinguished, however, by the spasms of the temporal and masseter muscles and of the tongue, also by the atrophy of the left half of the tongue and of the floor of the mouth under the tongue.

This exact association of symptoms has been observed in but one other case.[1] Spasms of the masseter and temporal muscles are not mentioned in the many records I have had access to, while atrophy of the tongue seems to be of more frequent occurrence.

The cases of progressive facial hemiatrophy present a great variety of symptoms. Any attempt at a physiological interpretation of the symptoms will reveal the fact that the disease cannot be limited to any one nerve. The old dispute, whether the trigeminus or the sympathetic be the chief sufferer may well be abandoned, for in some cases both are evidently involved; in some the trigeminus only, and in some the sympathetic only. It would be a simple matter, then, to disregard anatomical data and to make facial hemiatrophy a disease of trophic fibres — a true trophoneurosis. In addition to trophic fibres, motor fibres must be involved, or else the unusual

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[1] Axman and Hueter: Inaugural Dissertation. Berlin, 1848.

symptoms of my case—the motor spasm—could not be explained.

Trophic fibres, like their poor relations, the vasomotor fibres, have been a sort of medical bugaboo. Their existence seemed to be a postulate, yet no one felt able to prove that they were as real as motor fibres are. The physiologists denied their existence, while the pathologists had difficulty in explaining trophic symptoms except on the supposition of the existence of distinct trophic fibres. The trophic symptoms in cases of peripheral neuritis, in compression of the posterior root-fibres by tumors of the cord,[1] pointed indubitably to special trophic fibres in the peripheral nervous system. A little light has been thrown incidentally upon this subject in connection with the study of facial hemiatrophy.

Mendel[2] has been the only one[3] fortunate enough in recent years to obtain an autopsy in a well-defined case of this disease. The patient, who had passed through the hands of Romberg, Remak, Virchow, and others before she came to Mendel, had during more than twenty-five years presented all the symptoms of typical facial hemiatrophy complicated by atrophy of the left upper extremity. Erysipelas during confinement had immediately preceded the first onset of the symptoms. The autopsy, or, rather, the minute examination of the entire nervous system, revealed a neuritis interstitialis proliferans (Virchow) in all branches of the left fifth nerve, most marked, however, in the second branch; the facial was entirely normal, while the left musculo-spiral had undergone the same changes as the left trigeminal nerve.

Apart from some minor changes in the spinal cord (diminution of ganglion cells in the fifth cervical segment), which were secondary to the peripheral trouble, an atro-

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[1] Cf. the author's paper on Tumors of the Spinal Cord, Journal of Nervous and Mental Diseases, vol. xiii., No. 11, 1886.

[2] Neurologisches Centralblatt, 1888, No. 14.

[3] Jolly (Arch. f. Psych., vol. iii.) had an autopsy on a case complicated by multiple sclerosis.

phy of the descending root of the left fifth nerve, and a partial atrophy of the substantia ferruginea deserve most attention.

Mendel very justly claims that, as far as a single autopsy can prove anything, this one would show that the progressive hemiatrophy may be due to a peripheral neuritis (and in this he confirms a suspicion of Virchow), and furthermore it would indicate that the diseased parts, the descending root of the fifth nerve and the substantia ferruginea have trophic functions, presumably in connection with trophic fibres in the peripheral branches of the trigeminus.[1]

Referring once more to my own case, it is evident that the affection involves trophic as well as motor fibres of the trigeminal and hypoglossal nerves. It is a well-known fact that in some toxic cases of multiple neuritis the motor fibres of the peripheral nerves are more affected than the sensory. In some few cases the sensory fibres are involved long in advance of the motor fibres. There is, therefore, nothing foreign in the idea, if trophic fibres exist, that they be the seat of predilection for a disease which would seem to attack the peripheral nerves. My own case would show that this same affection may at times involve the motor as well as the trophic functions (fibres) of these nerves. In this connection it is worth noting that the disease often begins after some acute infectious disease, after erysipelas, and particularly after some traumatic injury to the face. And in these very conditions we recognize the etiological factors most frequent in well-known forms of peripheral neuritis. Lewin, in an excellent article on Hemiatrophy, has proved the traumatic history of 14 out of 68 cases. Mendel finds a peripheral origin in 19 out of 86 cases.

Facial hemiatrophy is evidently not related to the or-

[1] It is very evident that conditions of facial hemiatrophy may be of nuclear origin. Lowenfeld's case (cf. infra) points to this; the case of Flascher (Berl. kl. Wochenschr., 1880, No. 31), which Mills thinks peripheral, is possibly nuclear also.

dinary forms of muscular atrophy which have been the subject of so much discussion during the past years.[1] It would seem to hold kinship rather to other diseases in which the change in the nutrition of subcutaneous muscular and osseous tissue constitutes the chief feature. Lewin has described cases of hypertrophy of the face and body by way of contrast to these cases of hemiatrophy. Rosenthal has published a case in which the symptoms of facial hemiatrophy were combined with those of a partial scleroderma and alopecia areata, while Schulz, on the other hand, has published a case in which scleroderma was combined with Addison's disease and muscular atrophy. Tuczek [2] has reviewed these cases, together with that of Mendel, and has thus suggested an analogy between them, and it may be worth adding that the symptoms of facial hemiatrophy appear to be exactly the reverse of those which are presented in cases of akro-

[1] Mendel's case presented atrophy of the musculo-spiral muscles. In Cartwright's case mention is made of an atrophied spot in the deltoid. "The hole is about one inch in diameter, extending down to the bone, and can be felt through clothing"!

[2] Fortschritte der Medizin, December 1, 1889. For the general literature of the subject the reader is referred to: Eulenburg, Ziemssen's Cyclopedia, vol. xiv., 1877; Bannister, Journal of Nervous and Mental Diseases, vol. iii., 1876; G. Lewin, Charité-Annalen, 1884; Jessup and Browne, St. Bartholomew's Hospital Reports, 1882; C. K. Mills, Pepper's System of Medicine, vol. v.; Fürth, Lehrb. der Nerven-Krankheiten, p. 336.

NOTE.—In addition to the 71 cases collected by Lewin, and the 15 quoted by Mendel, I have been able to collect 11 others, bringing the total up to 97, including my own case. These eleven cases are the following: Eve, F. S., Transactions of the Pathological Society, London, 1887, vol. 38, p 81; Cartwright, H. P., Nashville Journal of Medicine and Surgery, 1888, N. S., vol. xli., p. 190; Mills, C. K.; Pepper's System of Medicine, vol. v., p. 694; Blumenau, L., Reviewed in Neurolog. Centralbl., January 1, 1890; Ephraim, Berl. kl. Wochenschr., 1889, No. 36 (reviewed in Neurol. Centralbl. No. 1, 1890); S. Rona, Reviewed in Neurolog., 1889, p. 216 (presented at a Budapest medical society); Lowenfeld, Münchener Med Wochenschr., 1888, Nos. 23 and 24 (nuclear case); Estor, Revue de Med, 1888, pp. 800 (complicated with hypertrophy of the upper lid, reviewed in Neurolog. Centralbl., 1889, p. 531; Ruheman, J., Centralblatt kl. Med., 1889, No. 1 (came on after operation for neurosis of upper jaw); Rosenthal, Berl. kl. Wochenschr., 1889, No. 34 (see text); Sachs, B., N. Y. MED. RECORD.

megaly; the symmetrical distribution and the general character of the symptoms in the latter disease, of course, forbidding any further inference as to the relation or differences between the two diseases. At all events, the peripheral nervous system in cases of akromegaly should be thoroughly examined post mortem.

In conclusion let me repeat that facial hemiatrophy is, in the main, a disease of the trophic nervous system, that the trigeminal and sympathetic nerves are the ones most frequently involved; but that the affection may spread to other cranial and peripheral nerves.